

COMMENTARY

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# Pulmonary arterial hypertension (PAH) related to congenital heart disease (CHD): introducing the CHAMPION supplement

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## Abstract

Early management decisions in complex congenital heart disease include assessing and maintaining adequate pulmonary blood flow whilst preventing increased pulmonary blood flow that may lead to pulmonary vascular remodelling and the development of pulmonary arterial hypertension. Such decision-making has now largely prevented the development of Eisenmenger syndrome in patients in the developed world, but there remain a large number of adult patients with congenital heart disease and pulmonary arterial hypertension (CHD-PAH).

The CHAMPION (Congenital Heart disease And pulMOnary arterial hyPerTension: Improving Outcomes through education and research Networks) group was formed to highlight the clinical needs of these patients and develop clinical research and education in this area, using a network approach. A highly attended and acclaimed educational event was held in late 2017, where experts in the field discussed the various aspects of CHD-PAH and presented cases in a variety of clinical scenarios, supported by facilitated discussion, with the aim of highlighting appropriate management strategies.

This supplement provides a detailed overview of CHD-PAH, including informative cases with discussion of the relevant management strategies.

**Keywords:** Pulmonary arterial hypertension, Congenital heart disease, Pulmonary vascular remodelling, Clinical management, Case studies

## Introduction

In 1897, Victor Eisenmenger described the case of a 32-year-old man who presented with cyanosis and evidence of right heart failure [1]. Within a year of presentation, the man had died following a large haemoptysis. Eisenmenger correctly attributed the presenting features to a large ventricular septal defect and hypothesized that pulmonary infarction led to the fatal haemoptysis. However, he attributed the cyanosis to poor peripheral circulation related to heart failure rather than to shunting.

“Eisenmenger’s case” was referred to by Abbott and Dawson in 1924, who reported a series of similar patients [2].

In 1958, Paul Wood, who was to influence so much in modern cardiology, used the term Eisenmenger syndrome (ES) to describe the process of an initial left-to-right shunt leading to increasing pulmonary vascular resistance and subsequent reversal of the shunt as the pulmonary arterial resistance exceeded that of the systemic circulation [3, 4]. He described all the conditions that we recognise as causes of ES, in a systematic description of the anatomy, physiology and natural history of each of these conditions. This characterisation, and the Heath Edwards classification, laid the foundation of modern care for all congenital heart disease.

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When managing any patient with congenital heart disease (CHD), it is important to be aware of their pulmonary blood flow and systemic saturations. With this knowledge, the management strategy varies depending on whether there is a balanced circulation or whether there is need to augment (by means of a systemic-pulmonary arterial shunt) or restrict pulmonary blood flow (pulmonary artery banding), to ensure that pulmonary arteries can grow whilst preventing the development of progressive pulmonary vascular remodelling. This approach requires significant expertise and paediatric cardiac services have been established in all developed countries to allow early recognition of CHD, which increasingly is identified in fetal life and has led to early corrective surgery in the majority of cases. Thus, where services are readily available the scenario rarely arises in which ES can develop, other than in association with a large atrial septal defect. ES remains a concern in parts of the world where such programmes do not exist.

There is an increased recognition that some patients will develop late pulmonary arterial hypertension (PAH) following corrective surgery and the reasons for this remain unclear. Late complications of corrective surgery include systemic ventricular dysfunction, valve disease and outflow tract obstruction and these can contribute to the development of pulmonary hypertension (PH) related to systemic ventricular dysfunction; however, these complications are more likely to result in postcapillary PH rather than PAH. There are also patients in whom PAH develops in the presence of a small left-to-right shunt, again for reasons that are not fully understood.

PAH related to congenital heart disease (CHD-PAH) is classified within Group 1 of the WHO classification of PH, and the categories described by Paul Wood are now recognised in a further classification of CHD-PAH, under four distinct categories (ES, left-to-right shunts, PAH with coincidental CHD and post-operative PAH) [5]. The introduction of oral targeted pulmonary vasodilator therapy for PAH has led to many patients receiving therapies with a positive impact on outcomes, both in terms of exercise capacity and pulmonary haemodynamics [6, 7], but also in terms of mortality when compared with historical controls [8]. It is important to correctly categorise patients with CHD-PAH into one of the four subgroups and, thus, decide on whether they should be offered PAH therapies. There remain significant challenges for patients with CHD-PAH and their clinical teams. For example, the use of standard echocardiography protocols for the diagnosis of PH [9] do not necessarily apply to patients with CHD where anatomical considerations, surgical pathways and certain physiologies pose challenges [10]. Expert care is essential for all CHD patients, especially those with suspected or confirmed PAH.

The investigation and management strategies for CHD-PAH patients are discussed in this supplement and illustrated with case studies. We discuss the existing evidence for the proposed strategies and recommended a consensus view where evidence is lacking. We address lifestyle issues that affect this patient group, including advice regarding contraception and pregnancy and approaches to minimise risks, and special circumstances, such as the management of non-cardiac emergencies, and the role of pulmonary vasodilators in the management of patients with a failing Fontan circulation.

The cases are presented as a practical guide to the management of this patient group, even though we recognise that an evidence-based approach is not possible in this setting. It is, therefore, beholden on clinical teams working in this area to develop collaborative clinical research programmes in order to enhance the evidence-base and improve care for these patients.

#### Abbreviations

CHD-PAH: Pulmonary arterial hypertension associated with congenital heart disease; ES: Eisenmenger syndrome; PH: Pulmonary hypertension; PAH: Pulmonary arterial hypertension

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